Modern Concepts of Cardiovascular Disease

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PULMONOCARDIAC FAILURE

Persons afflicted with severe deformities of the chest or spine are greatly handicapped throughout life, and, as a result of their deformity, it is unusual for them to reach old age. These crippled people often have back pain, chronic pulmonary infections, paraplegia or even esophageal obstruction. In addition to these complications many of them suffer with attacks which resemble sudden heart failure or asthma and it is these episodes which we have named Pulmonocardiac failure.[1] Our interest in this particular result of chest deformity was aroused by observing two persons who died in sudden attacks of pulmonocardiac failure.

The first was a man, 45 years of age, with severe kyphoscoliosis who had frequently recurring attacks of dyspnea and tachycardia for one year. His family doctor had attributed his symptoms to nervousness but the patient was so concerned that he had insisted on coming to the hospital. A diagnosis was made there of neurocirculatory asthenia and it was concluded that there was no structural cardiac defect. Five weeks later he was brought to the emergency ward in an attack resembling asthma and he died shortly after receiving morphia. The significant autopsy findings were the small, cramped lungs, hypertrophy of the right ventricle and evidence of congestive heart failure.

The second experience was with a young girl with severe kyphoscoliosis who complained of shortness of breath which was relieved by hyperextension in the recumbent position. This unusual method of obtaining relief suggested that her dyspnea was probably not due simply to heart failure. In confirmation of this a cardiac consultant could find no evidence for heart disease. Then a few weeks later, after a preoperative medication of pantopon, the patient suddenly developed extreme dyspnea and cyanosis and it was only through the prolonged administration of oxygen that she was saved. Within a year of this episode she returned to the hospital and died in a similar attack.

The dramatic end of these two patients so impressed us that we turned to the literature for an elucidation of what we had witnessed. Here we met with some disappointment as this bizarre syndrome was scarcely mentioned in the English medical literature. In contrast to this we did find numerous clinical notes on "scoliotic heart disease" in French and German. By 1883 Constantine Paul had included in his text book on heart disease an entire chapter entitled "Le Coeur de Bossus." But nowhere could we find an adequate explanation for the attacks nor could we find that anyone had attempted to study this aspect of the problem.

As the next step we discovered a number of young persons with chest deformities who were being followed in the Scoliosis Clinic of the Orthopedic department. Several of these complained of dyspnea and had various other symptoms including nocturnal smothering, asthma, fainting, palpitation, fatigue and weakness. Some of these unfortunates were considered to be psychoneurotic. Eight of these patients were taken to the Fatigue Laboratory at Harvard University and there, under the direction of Dr. D. B. Dill, we undertook a series of studies that were designed to estimate and explain the decrease in the functional capacity of their lungs and hearts.

The investigation began with determinations of body size, and lung volume and its subdivisions. Then the respiratory metabolism was studied by measuring the CO_2 production, oxygen utilization, respiratory volume, and the response of the subject to breathing a low oxygen gas mixture. In this last test about 12% oxygen was used which is the equivalent of attaining an altitude of 14,000 feet. In addition to routine observations on the circulation, determinations were made of the cardiac output, circulation time from arm to tongue and the venous pressure. Finally, various chemical properties of arterial blood were measured under normal conditions.

The most striking feature of our studies was the absolute and relative reduction in the lung volume. The values for vital capacity ranged from 700 cc. to 2520 cc., representing a reduction of over one-half normal in most instances. As this occurred, the ratio of residual air to vital capacity was found to be doubled. Consequent to this decrease in lung volume the respiratory minute volume was increased so that those with chest deformities have to perform more work to abstract the required amount of oxygen from inspired air. This was clearly shown by the fact that those with chest deformities breathe 25 to 35 liters of air for each liter of oxygen absorbed, while the normal twin brother of one of the subjects used only 22 liters.

In testing the response to low oxygen tensions one subject caused considerable alarm by having an acute attack of pulmonocardiac failure. She became cyanotic, pulseless, and lost consciousness after breathing a 12% oxygen mixture for a few minutes. Her recovery was very gradual but when a second patient had a similar attack following the test we desisted from further such studies. These incidents demonstrated to us the unfitness of these persons, which is due, in large part, to pulmonary insuffi-

1. The Decrease in Functional Capacity of the Lungs and Heart Resulting from Deformities of the Chest: Pulmonocardiac Failure. Earle M. Chapman, D. B. Dill and Ashton Graybiel. *Medicine* 18, 167, 1939.

ciency. Additional evidence that the chief effect of this syndrome is on the lungs is found in the clinical facts that respiratory depressants, pulmonary infection, a cramped position, increase in the deformity or any process that further reduces pulmonary function may lead to frank pulmonocardiac failure. It is likely that in severe funnel chests the mediastinal displacement and rotation are also important factors.

Although exact measurements of the pressure in the pulmonary artery are not feasible we concluded that this pressure is usually increased. This would afford an explanation for the hypertrophy and dilatation of the right ventricle, frequently seen at necropsy, and the accentuation of the second pulmonic sound often heard on physical examination. Pulmonary hypertension, with its well-known effect of retarding the coronary circulation of the right ventricle, may be an important contributory factor in precipitating the attacks of pulmonocardiac failure we have observed. We found, at a time when our subjects were not in congestive failure, that the circulation time through the lungs was normal, that the venous pressure was normal, and that the arterial pressures and cardiac output are usually normal.

The only definite electrocardiographic abnormality was the occasional occurrence of right axis deviation.

We were fortunate in having the normal twin brother of one hunch-backed man for comparative study and from this we substantiated our clinical impression that the general development of the body is greatly retarded by spinal curvature.

From our studies of these cases and from a review of the literature, it became apparent that we were dealing with a true clinical syndrome. The succession of pathological changes in the lungs and heart follows a recognizable course which in turn provokes symptoms of a definite clinical pattern, which should be recognized by the surgeon, the internist and particularly the anesthetist.

From all sources we were able to find 126 reported fatal cases of this syndrome and to this we have added four cases. The average age at death of 79 patients was 30 years. For some unaccountable reason the incidence of right-sided scoliosis is far greater than left. This may be significant in that the secondary or compensatory curve of the spine compresses the left chest.

Dyspnea is the chief complaint of these people. Because of the distortion and rigidity of the thoracic cage, they breathe with difficulty and often rely almost entirely upon the diaphragm for the necessary movement. This leads to an habitual state of dyspnea which increases gradually after the arrest of skeletal growth about the age of 20. Thereafter, with exertion or because of some factor affecting pulmonary efficiency, they may become subject to attacks of paroxysmal dyspnea and episodes of great weakness and fainting. This transition from habitual dyspnea to the more severe symptoms marks the onset of pulmonocardiac failure. Once severe symptoms appear the interval before death is usually short. The rapid development of pulmonocardiac failure gives little advance warning to the physician. Precordial pain is a rare complaint and so the confusion with myocardial infarction from coronary occlusion is unlikely although we have seen such an error in diagnosis in an additional case recently under observation.

The chief signs of this combined failure of the lungs and heart are dyspnea and a persistently rapid heart rate. The heart sounds may be normal or a bit distant and accentuation of the second pulmonic sound is usually present. Changes in the heart size and shape are difficult to detect in the presence of a chest deformity but right-sided cardiac enlargement may be seen by fluoroscopy. Hypertension is seldom present but an evelation of the diastolic pressure may often cause a small pulse pressure.

The morbid anatomy has been adequately described in 69 of the cases reviewed and in one of our own cases. Forty-nine of these 70 had definite enlargement of the right ventricle. The changes in the lungs have been variable and included pneumonitis, bronchitis, atelectasis, bronchiectasis and emphysema. Most often a decrease in actual lung substance was noted. Loss of lung surface in a compressed area was seldom offset by the compensatory emphysema in the region less restricted by the ribs. Our own explanation of this difference in observations in regard to the pulmonary changes is that the type of pulmonary change depends on the age at which the deformity was acquired. Before the age of 18 to 20 years, chest deformity may prevent the lungs from developing to their full size, hence one would find a cramped organ that would resemble a child's lung. After this age of full skeletal growth one can expect atrophy in the compressed region and emphysema in the region of greater function. This idea is supported by the work of Bremer who has shown that the normal lung grows by an increase in the number of alveoli and not by an increase in their size. After lobectomy he found actual regeneration of lung tissue occurring in the young while only dilatation of the alveoli occurred in the adult whose lungs had stopped growing.

This syndrome of pulmonocardiac failure is not the same as cor pulmonale or Ayerza's disease. It stands alone in its peculiar manifestations.

Treatment of the chest deformities is obviously handicapped by the structural changes in the skeleton. Every effort to relieve the deformity should be pursued and supportive measures which embrace a position of hyperextension should be tried. The orthopedic surgeons should work in the closest cooperation with the internist and the anesthetist in preparing these subjects for such procedures as spinal fusion, rib resection or other measures which may alleviate them. The patient should be encouraged to breathe deeply and to pursue exercises such as using blow bottles to increase the vital capacity. Certainly a precarious level is approached when the vital capacity drops below 1200 cc. Activity should be limited and exercise at high altitudes or plane travel should be forbidden. The prevention of respiratory tract infection is of the greatest importance and respiratory depressants and anesthetics are to be used with considerable caution. In the event of actual heart failure little aid can be expected from the remedies usually employed. Bed rest in hyperextension is a surprising form of relief for some of those with pulmonocardiac failure.

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